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# Synopses

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# Dental Implications of Chronic Renal Disease in Children

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## Introduction

The estimated incidence of end-stage renal failure (ESFR) in childhood due to either congenital or acquired conditions is 10-12 per 1 million children, with a prevalence that ranges between 39-56 million children. Significant improvement in dialysis treatments as well as outcomes of renal transplantation has led to more children and adolescents living longer with renal disease (1).

Since chronic renal disease is characterized by multi-organ involvement, dental clinicians, especially paediatric dentists, are likely to manage an increasing number of children and adolescents with oral manifestations associated with renal disease. This article provides a review of the anatomy and functions of the kidney, the pathophysiology of chronic renal disease, oral manifestations and their clinical implications.

## Anatomy and Function of Kidney

Kidneys have multiple functions, especially in fluid and electrolyte balance, waste removal, acid-base balance, vitamin D metabolism and stimulation of erythrocyte production. Kidneys produce urine by filtering small molecules and ions from the blood and then salvaging the useful materials such as glucose. The nephron is the basic structural unit of the kidney and comprises a Bowman's capsule, glomerulus (a capillary network within Bowman's capsule), proximal convoluted tubule, loop of Henle, and distal convoluted and collecting tubules (2).

The kidneys pass urine through the ureters to the bladder where it is stored and eventually passed through the urethra. Approximately a quarter of blood volume perfuses the kidney each minute. The kidneys are involved in the excretion of drugs and hormones and also act as endocrine organs for the production of hydroxycholecalciferol (active vitamin D), erythropoietin, renin, prostaglandins, and as target organs for the parathyroid hormone and aldosterone. Renal failure can lead to fluid retention, acidosis, accumulation of metabolites and drugs, damage to platelets (leading to bleeding tendency), hypertension, anaemia and endocrine effects (2).

Renal diseases can be classified as acute, chronic, acquired or congenital conditions (3). From birth to ages 4, congenital defects and hereditary disorders are the leading causes of kidney failure. Between the ages range of 5 and 14, other causes such as immune, inflammatory and infectious conditions play a more significant role. (4).

## Pathophysiology of Chronic Kidney Disease

Chronic kidney disease (CKD) is defined as kidney damage or a reduction in the glomerular filtration rate (GFR) for 3 or more months. Individuals with a GFR of less than 90ml/min per 1.73m2 for 3 months in association with proteinuria or haematuria are classified as having CKD. These patients are at an increased risk of loss of kidney function and subsequent development of cardiovascular disease (2).

The aetiology of CKD can be classified according to the part of renal anatomy that is affected. The disease can involve the larger vascular pedicle (e.g. bilateral renal artery stenosis) or the microvasculature (e.g. ischaemic nephropathy, vasculitis). The pathology may also involve the glomerulus and these can be categorized into primary (e.g. focal segmental glomerulosclerosis, immunoglobulin A nephritis) or secondary (e.g. diabetic nephropathy, lupus nephritis) diseases. Diseases also affect the tubolointerstitial segment of the glomerulus. These include polycystic kidney disease, drug – and toxin-induced chronic

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## THIS ISSUE

# Colgate® Corner

*by Sue Cartwright,*
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## caries? we care!

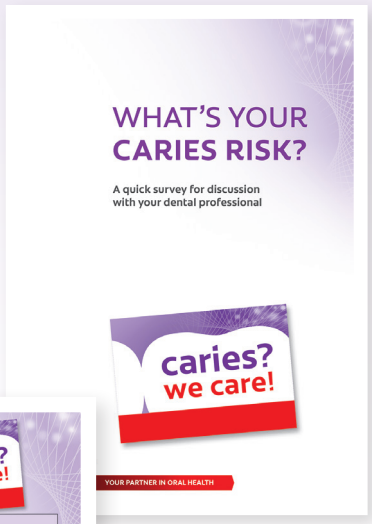
Prevention of dental caries is a goal many practitioners strive towards. It is very hard to achieve with any sort of certainty. It may involve changing the way you practise and this is indeed a challenge. It will almost certainly involve the patient in changing some of their habits, which is also a difficult task.

We would like to help you with this process through the provision of materials based on the International Caries Classification and Management System (ICCMS). This system grew out of work done by a global collaboration of cariologists and uses the International Caries Detection and Assessment System (ICDAS) which you can find at **www.icdas.org**. There are training materials and guidebooks available on this site which will help you with the implementation of a comprehensive caries management program in your practice.

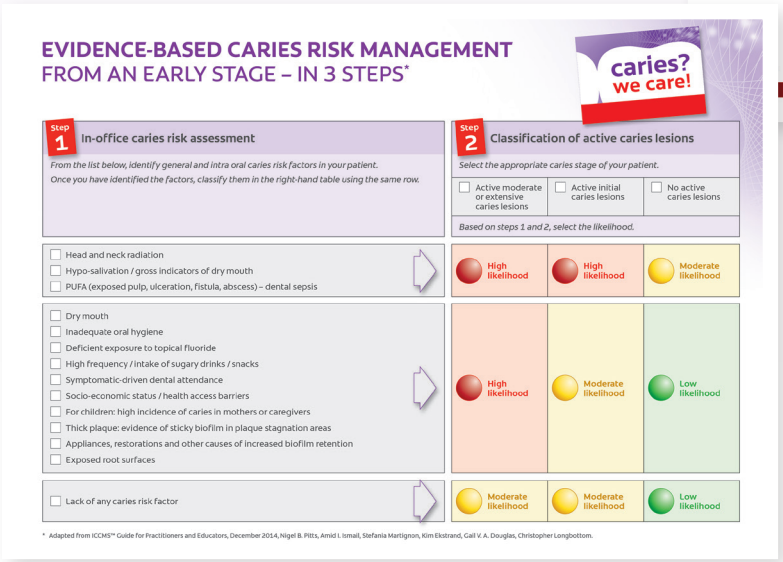
The materials we can offer you will help with risk assessment, diagnosis and management strategies based on ICCMS as well as providing the basis for a discussion with the patient and messages that can be sent home with them.

The caries? we care! materials are available as writable PDFs which can be downloaded/printed from the Colgate Professional website at **www.colgateprofessional.com.au or www.cogateprofessional.co.nz**.


RISK EVOLUTION CARD




PATIENT LEAFLET




CHAIRSIDE MATERIALS

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tubolointerstitial nephritis and reflux nephropathy. Finally obstructive pathology can also cause disease. The most common of these causes include renal or bladder stones and prostate diseases (2).

Chronic kidney disease occurs when the damaged kidneys have reduced function leading to waste substances like urea to accumulate (2).

Risk factors of CKD include cardiovascular disease, obesity, hypercholesterolaemia and a family history of CKD. Poorly controlled diabetes and hypertension can increase the risk of progression of CKD to kidney failure. Recurrent kidney injury such as in cases of infection, drugs or toxins injurious to the kidney can lead to progression for kidney failure especially in older adults. CKD is usually irreversible and progressive and can lead to end-stage renal disease (ESDR). CKD can be compensated by structural and functional hypertrophy of surviving nephrons until 50% of renal function remains. However with continued progression, chronic renal insufficiency will ensue (2). The five stages of CKD according to the National Kidney Foundation are summarized in ***Table 1***.

Patients with early CKD often have no symptoms, with only a blood test showing diminished kidney function and a urine test to assess kidney damage. When kidney function has fallen to less than 25% of normal, symptoms such as nocturia and anorexia occur as well as increased serum levels of nitrogenous compounds (i.e. urea). Later problems include cardiovascular disease (heart attacks, heart failure, heart rhythm disturbances and strokes), anaemia and bone disease (2).

## General Management

CKD is diagnosed by clinical history and presentation and by increasing levels of plasma urea and creatinine as well as decreasing levels of estimated GFR. The reduction in eGFR can be plotted over time allowing clinicians to predict prognosis accurately and plan for treatment such as timing of dialysis (2).

The primary causes of CKD should be treated where possible and any stress, infection or urinary obstruction should be managed as they may precipitate symptoms. The aim of treatment is to slow down or halt the progression of CKD to Stage 5 as well as decrease the risk of cardiovascular disease, which is the main cause of mortality. A normal diet, potassium restriction and salt or water control are utilized. As patients with a low GFR have a high risk of cardiovascular disease, management involves use of statins, aspirin in conjunction with smoking cessation and other measures (2).

Symptoms and complications such as vomiting, fits and calcium loss are also treated. Iron levels are maintained by using iron and epoetin alfa recombinant (recombinant erythropoietin) or darbepoetin alfa, which has a longer half-life. Calcium carbonate, vitamin D3 or its synthetic analogue, a low-phosphate diet or intravenous sodium clodronate assists to inhibit bone resorption. Cinacalcet, which reduces parathyroid hormone, may also be used. A parathyroidectomy may be required in cases of tertiary hyperparathyroidism while a bisphosphonate may be taken for the management of renal osteodystrophy (2).

Drug therapy needs to be carried out with caution as dosage of drugs cleared renally (e.g. penicillins, cephalosporin and erythromycin) should be reduced according to renal function. The least nephrotoxic agents should be utilized (i.e. NSAIDS or tetracyclines should be avoided) and alternative medications should be considered to avoid potential drug interactions (2).

CKD can be managed through medication and lifestyle changes to slow disease progression and to prevent or delay the onset of kidney failure. However, the only treatment for Stage 5 of CKD (i.e. end-stage renal failure) is renal replacement therapy- dialysis or transplantation (2).

### *Dialysis:*

There are two main types of dialysis – peritoneal dialysis and haemodialysis. In both situations, the patient's blood is separated from the dialysis fluid (dialysate) by a membrane, which allows water and toxins, but not blood cells to pass out of the blood (2).

### *Peritoneal dialysis:*

Peritoneal dialysis works on the principle that the peritoneal membrane (a serous membrane that forms the lining of the abdominal cavity) can act as a natural semi-permeable membrane. Dialysis fluid is introduced via a catheter placed near the umbilicus into the abdominal cavity or tunnelled under the skin near the sternum. Advantages of peritoneal dialysis include it being relatively easy to learn, fluid balance is easier than with haemodialysis and it's usually done at home. Even though it is less efficient than haemodialysis, it is as beneficial overall since it is performed more frequently (2).

Continuous ambulatory peritoneal dialysis (CAPD) has about 4-5 manual changes daily while continuous cyclic peritoneal dialysis (CCPD) is a machine that does the exchanges at night. Intermittent peritoneal dialysis (IPD) uses the same type of machine as CCPD and if done overnight is call nocturnal intermittent peritoneal dialysis (NIPD). Control of infection is the key with peritoneal dialysis carrying the risk of peritonitis (as well as fluid leaks into adjacent tissues and hernias). Infections of the PD catheter exit site or 'tunnel' are less serious (2). Peritoneal dialysis is preferred in children, with some advantages being fe wer dietary restrictions, better growth, less bone disease and anaemia and it also allows the child to have a relatively normal life with less frequent hospital visits when compared with haemodialysis (1).

### *Haemodialysis:*

Haemodialysis is used to remove metabolites such as urea, potassium and excess water by exposing the patient's blood to a hypotonic solution across a semi-permeable membrane, which allows diffusion and osmosis of solutes and fluid from the body. The dialysis solution has concentrations of

| *Stages* | *Renal Health* | *GFR mL/min/1.73m2* | *Features* |
|---|---|---|---|
| | Normal | 130 | |
| 1 | Diminished renal reserve (early CKD) | >90 | Abnormalities in blood or urine tests or imaging studies but few overt symptoms |
| 2 | Mild CKD (azotaemia) | 60-89 | Abnormalities in blood or urine tests or imaging studies |
| 3 | Moderately severe | 30-59 | Abnormalities in blood or urine tests or imaging studies |
| 4 | Severe CKD | 15-29 | Uremic symptoms |
| 5 | End-stage renal failure (ESFR) | <15 | Life-threatening and requires some form of renal replacement therapy |

*Table 1. Stages of Chronic Kidney Disease (5)*

minerals such as potassium and calcium that are similar to their natural concentration in healthy blood. For other solutes such as bicarbonate, the dialysis solution level is set at a slightly higher level than normal blood concentrations to encourage diffusion of bicarbonate into the body and hence neutralise the metabolic acidosis that is commonly present (2).

Haemodialysis is often carried out at home or as an outpatient. An arteriovenous fistula is usually formed surgically above the wrist to allow the introduction of infusion lines. Alternatively, a Gortex® or a PTFE graft is placed or an indwelling, tunneled cuffed catheter can be used. The patient is heparinised during dialysis to keep both the infusion lines and the dialysis machine tubing patent and the patient's blood is passed through an extracorporeal circulation (2).

*Haemofiltration:*

In haemofiltration, blood is pumped through a "haemofilter" such as in dialysis however instead of a dialysate (a component of the mixture which passes through the membrane dialysis) being used, a pressure gradient is applied. Water therefore moves rapidly across the very permeable membrane assisting the removal of metabolites, especially those with large molecular weights that are cleared less well by haemodialysis. The ions and water that are lost from the blood during the process are replaced with a "substitution fluid" which is infused into the extracorporeal circuit during treatment. Haemodiafiltration combines haemodialysis and haemofiltration into one process (2).

Dialysis is able to totally rehabilitate up to 20% of patients, however, it cannot prevent all complications. Adverse effects of the procedure (collectively termed as a dialysis "hangover" or "washout") is caused by removing fluid too rapidly or excessively and can include symptoms such as hypotension, cramps, febrile reaction, arrhythmia and hypoxaemia. Long-term adverse effects include ischaemic heart disease, cardiac valve calcification (especially affecting the aortic valve), dialysis-related amyloidosis and neuropathies. Cardiovascular disease and infection are the most frequent causes of death. Additionally, haemodialysis may damage platelets mechanically, which will further exacerbate a bleeding tendency (2).

Patients with HD grafts and catheters may be at risk of infection and bacterial endocarditis, osteomyelitis, blood-borne virus infections (i.e. hepatitis or HIV) and tuberculosis, which were common in the past (2).

*Renal transplant:*

Renal transplantation is now the gold-standard treatment choice for children with end-stage renal disease, as it allows them a better quality of life and increases their long-term survival compared to any other form of renal replacement therapies (6,7). Generally, the outcome of renal transplantation in children with young adult donors is significantly better than in adults. One of the main factors that determine the success of the transplant is the compatibility of the antigens associated with blood type (ABO system) and human leucocytes antigens (HLA). Hence before the transplant is performed, the blood or tissue of the donor and recipient is matched to at least prevent hyper-acute rejection due to pre-formed antibodies between the donor and the recipient. Immediately pre-transplantation and thereafter, the child will require immunosuppressive treatment, which now usually consists of corticosteroids, a calcineuri-inhibitor such as cyclosporine or tacrolimus and a lymphocyte-proliferation inhibitor such as Azathioprine. Renal transplant survival rates after cadaver-donor kidney grafting can now reach 83% after 1-year and 65% after 5-years. Survival after transplantation from living donors has been reported to be approximately 10-15% greater (1).

## Dental Implications of Chronic Renal Disease

The most common oral manifestation of chronic renal disease or to its treatment include gingival enlargements, periodontal disease, susceptibility to dental caries, salivary changes, halitosis/altered taste sensation, oral soft tissue changes, oral malignancies, dental anomalies and bone metabolism.

*Gingival enlargements:*

The gingivae in patients affected with chronic renal failure can be pale due to anemia with possible loss of the demarcation of the muco-gingival junction. However, this has now been nearly eliminated by management of anaemia with erythropoietin. Patients can also suffer from bleeding, petechiae and ecchymosis due to uraemia. Gingival bleeding can also be exacerbated by platelet dysfunction as well as patients being on anti-coagulants for dialysis purposes (1, 6).

Gingival hyperplasia is prevalent in patients with organ transplant and is clinically highly variable. It severity is dependent on several factors including age (with children experiencing at a more prevalent rate), gender (males greater than females), the severity of the underlying condition, degree of immunosuppression, phamarcokinetic properties of the medication especially the immunosuppressants, the presence of gingivitis before transplantation, individual susceptibility to the medication and their genetic predisposition (1,6).

Initially, gingival hyperplasia is managed conservatively with meticulous professional and personal oral hygiene. This will assist in reducing the inflammatory component of the gingival overgrowth with an improvement in oral hygiene leading to a reduction in gingival overgrowth in patients with continuous cyclosporine therapy. Treatment such as surgical reduction by laser or traditional surgery can be considered, however, there is a high-risk of recurrence (1,6). Chabria et al. (8) recommended a delay in repeat surgery for at least 3-years to reduce the recurrence rate. Regardless, it is crucial to establish a meticulous oral hygiene program for children receiving immunosuppressants such as cyclosporine or calcium-channel blockers in order to reduce the side-effects of these medicaments on the gingivae (8).

*Periodontal disease:*

The oral hygiene of patients receiving haemodialysis can be poor. A study by Naugle et al. (9) showed only 15% of 45 patients receiving haemodialysis had good oral hygiene standards. Calculus deposits may also be increased due to high salivary urea and phosphate levels. Other important risk factor for the development of dental calculus and dark brown staining of teeth is the ingestion of large quantities of calcium carbonate. Extrinsic staining of teeth is most likely due to intake of liquid ferrous sulphate therapy, which is given for the management of anaemia (10).

At present, studies show conflicting data regarding periodontal disease. There is no evidence showing patients are at a higher risk of periodontitis, however, premature loss of teeth has been reported (6). It is believed that cases of severe periodontitis in patients with renal disease and those with associated polymorphonuclear leukocytes (PMN) impairment may develop increased susceptibility in those that do not access early treatment (1).

*Salivary Changes:*

Patients with chronic renal disease commonly experience symptoms of xerostomia, especially in individuals

receiving haemodialysis. Possible causes include restricted fluid intake, side-effects of drug therapy and/or mouth breathing. Long-term xerostomia can predispose the patient to caries and gingival inflammation. This can lead to difficulties with speech, denture retention, mastication, dysphagia, sore mouth and loss of taste. It also increases the risk caries and infections such as candidosis and acute suppurative sialadenitis (6).

A study by Al Nowaiser et al. (11) reported that children with chronic renal disease have significantly more alkaline salivary pH compared to a group of renal transplant children. This is thought to be due to the presence of salivary proteins, potassium and sodium at greater concentrations (12).

*Halitosis/Altered taste sensation:*

Uraemic patients may have an ammonia-like oral odour, which can occur in approximately 1/3rd of individuals receiving haemodialysis. Chronic renal failure can also produce altered taste sensations and some patients complain of an unpleasant and/or metallic taste or a sensation of an enlarged tongue (6).

*Oral soft tissue changes:*

Wide ranges of oral mucosal lesions have been described especially white patches and/or ulcerations in patients receiving dialysis and allografts. Lichen-planus like disease can sometimes be due to associated drug therapy such as the use of diuretics and beta-blockers. Oral hairy leukoplakia has also been known to occur secondary to drug-related immunosuppression (6).

Uremic stomatitis can manifest as white, red or grey areas of the oral mucosa. The erythemapultaceous form can consists of a grey pseudomembrane overlying painful erythematous patches. The ulcerative form can appear red with a 'pultaceous' covering. There are no good histological description of uremic stomatitis hence it is difficult to define the cause of this unusual oral mucosal change. Uraemic stomatitis has, however, been thought to be due to chemically-based trauma from increased levels of nitrogenous compounds (6).

In some patients, the mucosal surface may become erythematous or ulcerate. Oral mucosal macules and nodules have been reported in 14% of patients receiving haemodialysis. Other lesions that can occur intra-orally include fibro-epithelial polyps, geographic tongue, black hairy tongue, papilloma and pyogenic granuloma. Angular cheilitis have been noted in up to 4% of haemodialysis and renal allograft patient (6).

*Oral Malignancy*

The risk of oral squamous cell carcinoma in patients undergoing haemodialysis is generally similar to that of otherwise healthy patients in the general population. Kaposi's sarcoma and non-Hodgkin's lymphoma have, however, been reported to occur at a higher rate which is most likely due to the iatrogenic immunosuppression in patients with chronic renal failure, which increases their risk to these virally-associated tumours (6).

*Dental Anomalies*

Enamel hypoplasia of the primary and permanent teeth with or without brown discolouration is frequently seen in patients with renal disease and is attributed to the production of thin enamel due to disruption of ameloblast function during enamel matrix secretion. Factors responsible for this disturbance include hypocalcaemia, decreased serum levels of 1, 25-dihydroxycholecalciferol and increased serum levels of inorganic phosphate and serum parathyroid hormone. A study by Scheutzel et al. (13) observed enamel hypoplasia in 52% of patients with chronic renal failure.

Delayed eruption of permanent teeth has also been reported in children with chronic renal failure as well as narrowing or calcification of the pulp chambers of teeth in adults. The cause of this narrowing is unknown. Renal allograft patients have significantly more narrowing of the pulp chambers to those patients on haemodialysis. No consistent association between the use of corticosteroid therapy and narrowing of the pulp chamber has been reported (6). It is believed to be more likely due to a disturbance of calcium and phosphate metabolism (14,15)

*Dental caries:*

There is no evidence of a significantly increased risk of dental caries in patients with renal failure. Generally the prevalence of dental caries has been reported to be low due to increased pH levels of the saliva as a result of increased concentrations of salivary urea (12). A study by Al Nowaiser et al. (16) found that children with chronic renal failure experienced 30% less caries compared to healthy children. Non-carious tooth loss is, however, more prevalent in patients with CRF than the general population. This may be due to nausea, oesophageal regurgitation or induced vomiting in bulimia nervosa (especially if the patient finds the restricted diet unpleasant) (6).

*Bone Lesions*

A wide range of bony anomalies can occur due to chronic renal disease. This occurs due to defects in calcium metabolism including loss of hydroxylation of 1-hydroxycholecalciferol to activate vitamin D (1,25 dihydroxycholeccalciferol), decreased hydrogen ion excretion and resultant acidosis, hyperphosphataemia,

| *Bony Features* | *Features of Teeth and Periodontium* |
|---|---|
| Bone Demineralization | Delayed eruption |
| Reduced trabeculation | Enamel hypoplasia |
| Reduced thickness of cortical bone | Loss of the lamina dura |
| Ground-glass appearance of bone | Widening of the periodontal ligament |
| Metastatic soft-tissue calcifications | Severe periodontal destruction |
| Radiolucency fibrocystic lesions | Tooth mobility |
| Radiolucency giant cell lesions | Drifting |
| Lytic areas of bone | Pulp calcifications |
| Fractures of the jaw as a results of trauma or surgery | Pulp narrowing |
| Abnormal bone healing post-extractio | |

*Table 2. Orofacial features of renal osteodystrophy (6)*

hypocalcaemia with resultant secondary hyperparathyroidism and interference in phosphate biochemistry by dialysis (6).

Secondary hyperparathyroidism can affect up to 92% of patients undergoing haemodialysis. Hyperparathyroidism can present as a maxillary brown tumour, enlargement of the skeletal bases or tooth mobility (6). Parathyroid hyperplasia may eventually become adenomatous and irreversible (tertiary hyperparathyroidism) (2). Orofacial features of renal osteodystrophy due to hyperparathyroidism are summarized in ***Table 2***.

## Implications in Dental Management of Children with Chronic Kidney Disease

Untreated dental infections in immunosuppressed patients can lead to morbidity and transplant rejection. Hence, regular dental reviews are important for early detection of oral disease. Clinicians before treatment must liase with paediatric nephrologists to manage possible complications related to bleeding tendencies, hypertension, risk of adrenal crisis, susceptibility to infection, anemia and drug clearance

### *Bleeding tendency:*

Bleeding tendency is key clinical feature in patients with chronic kidney disease, which have serious implications to dentistry. Bleeding tendencies can occurs in CKD because of diminished thrombopoietin production and defective von Willebrand factor. This causes a cascade of events that results in abnormal platelet production and function. Additionally, a reduction in platelet factor 3 impairs the coagulation pathway by inhibiting the conversion of prothrombin to thrombin, which in turn will further exacerbate bleeding (2). Patients undergoing haemodialysis also have a bleeding tendency as a result of anti-coagulants or platelet dysfunction (6).

If a dental procedure with a risk of bleeding is to be performed, a haematologist should be consulted. Dental treatment is usually best carried out on the day after dialysis where there is the greatest benefit from the dialysis and the effect of the heparin has subsided. Haemostasis should be ensured once surgical procedures are complete. If bleeding is prolonged, desmopressin (DDAVP) may provide haemostasis for up to 4 hours. If this fails, cryoprecipitate may be effective, however, its peak effect is at 4 – 12 hours and lasts up to 36 hours (2).

### *Hypertension:*

Arterial hypertension is a common finding in children through all stages of CKD. It is due to a combination of factors including fluid overload and activation of the renin-angiotensin system, sympathetic hyperactivation, endothelial dysfunction and chronic hyperparathyroidism. Additionally several drugs taken regularly by children with CKD such as erythropoietin, glucocorticoids and cyclosporine A are known to independently increase blood pressure in a dose-dependent manner. Blood pressure is one of the key determinants of progression rate of renal failure as well as cardiovascular mortality in children with renal failure (17).

For high-risk patients, blood pressure should be monitored for white-coat hypertension, which may be triggered by anxiety from the dental visit. For patients taking anti-hypertensive medication such as calcium-channel blockers, gingival hyperplasia is a common side effect. Xerostomia has also been associated with anti-hypertensive treatment, mainly in adults. Clinicians must also be cautious about the interactions between non-steroidal anti–inflammatory medications (NSAIDs) and ACE inhibitors, beta-adrenergic blockers and diuretics. Their use should be limited in order to prevent reducing the anti-hypertensive effects of these drugs. For children with poorly managed hypertension, use of local anesthetics with vasoconstrictors should be avoided (18).

### *Risk of adrenal crisis:*

To reduce the risk of adrenal crisis in patients who have taken large doses of corticosteroid (10 mg of prednisolone daily or equivalent during the preceding 3 months) and who are undergoing invasive surgical procedures such as extractions of more than one tooth, appropriate corticosteroid cover should be administered. The nephrologist must be consulted regarding the need and dosage (6).

### *Susceptibility for Infection and Antibiotic prophylaxis:*

Infections are poorly controlled in patients with CKD especially if immunosuppressed. Patients with CKD can be immunosuppressed due to defective phagocytic function as a result of decreased interleukin-2 (IL-2) and increased levels of pro-inflammatory cytokines (IL-1, IL-6 and tumour necrosis factor- TNF) (2). Patients with renal transplants are affected by drug-related immunosuppression.

Infections in these patients may spread locally as well as initiating septicaemia. They can also accelerate tissue catabolism leading to clinical deterioration with some evidence showing periodontitis can perpetuate inflammation in CKD (2). These infections can be difficult to recognise in immunocompromised patients as the signs of inflammation are masked. Odontogenic infections should be treated vigorously i.e. extraction (2). Vascular access infections are generally caused by skin organisms such as Staphylococcus aureus and are only rarely affected by oral microorganisms. Hence, patients with most arteriovenous fistulas or indwelling intra-peritoneal catheters are not considered to require antimicrobial prophylaxis (2).

Patients with a compromised immune system may not be able to tolerate transient bacteraemia following invasive dental procedures. These non-cardiac factors are able to place a patient with compromised immunity at risk for distant-site infections from a dental procedure. However, different countries have varying guidelines regarding antibiotic prophylaxis. The British Society for Antimicrobial Chemotherapy (BSAC) guidelines does not recommend antibiotic prophylaxis for individuals with renal disease who require invasive dental procedure. In contrast the 2014 American Academy of Pediatric Dentistry (AAPD) guidelines, which follow the 2007 American Heart Association (AHA) guidelines and is also supported by the Australian Therapeutic 2012 guidelines, recommend consultation with child's nephrologist regarding antibiotic prophylaxis for high-risk dental procedures in immuno-compromised children (19). Antibiotic prophylaxis, in consultation with the nephrologist, is also recommended in patients with renal transplants, polycystic kidney disease type I with mitral valve prolapse and mitral valve regurgitation, those on peritoneal dialysis or patients with prosthetic bridge grafts or PTFE or tunnelled cuffed catheters due to risk infection of the vascular access site (2, 6, 20).

Klassen and Krasko (21) have highlighted the importance of having good oral health for patients with chronic renal disease as it lowers their risk of septicaemia, endocarditis or endoarteritis at the site of vascular dialysis.

*Table 3. Commonly used drugs in dentistry and their implications for patients with renal disease (6)*

| *Drug* | *Caution* |
|---|---|
| ANTIBIOTICS | |
| Amoxicillin, ampicillin | Reduce dose, rash more common. |
| Erythromycin | Maximum 1.5g daily (ototxicity). Increases plasma-tacrolimus concentration. Increases plasma-cyclosporin concentration. |
| Tetracyclines | Avoid – use doxycycline or minocycline if necessary (avoid excessive doses). Doxycycline increases plasma-cyclosporin concentration. |
| Cefalexin, cefradine | Reduce dose. |
| Probenecid | Avoid (ineffective, increased toxicity). |
| ANTIFUNGALS | |
| Amphotericin | Use only if no alternative. Cyclosporin, tacrolimus: increase risk of nephrotoxicity. |
| Fluconazole | Usual initial dose then halve subsequent doses. Increases plasma-cyclosporin concentration. Increases plasma-tacrolimus concentration. |
| Miconazole | Increases plasma-cyclosporin concentration. |
| ANTIVIRALS | |
| Acyclovir | Reduce dose. |
| ANALGESICS | |
| Aspirin | Avoid (sodium, water retention; deterioration in renal function; risk of gastric hemorrhage) |
| Ibrofen, diflunisal | Avoid if possible/use lowest effective dose, monitor renal function (sodium, water retention; deterioration in renal function). Cyclosporin, tacrolimus: increase risk of nephtrotoxicity. |
| Dihydrocodeine, pethidine | Reduce dose/avoid (increased and prolonged effect, increased cerebral sensitivity). |
| OTHER DRUGS | |
| Carbamazepine | Caution. Reduces plasma-cyclosporin concentration. |
| Nitrazepam, temazepam | Start with small doses (increased cerebral sensitivity). |
| Povidone-iodine | Avoid regular application to inflamed or broken mucosa. |
| Ephedrine | Avoid (CNS toxicity) |

*Affect on Drug Metabolism and Dosage:*

Drugs that are excreted mainly from the kidneys may have enhanced or prolonged activity if doses are not lowered. This is mainly dependent on factors such as the degree of renal failure, the dialysis schedule or the presence of a transplant. Hence, it is imperative that drugs should be prescribed only after consultation with the renal consultant (2, 22).

Drugs that are directly nephrotoxic must be avoided. Aspirin (which already may be given as a prophylaxis against cardiovascular disease) and other NSAIDs should be avoided as they aggravate gastrointestinal irritation and bleeding associated with CKD. Their excretion may also be delayed and they may be nephrotoxic, especially in older patients or where there is renal damage or cardiac failure. Use of NSAIDs is associated with risk of acute renal failure, nephrotic syndrome with interstitial nephritis and CKD. They also reduce sodium excretion, which can lead to peripheral oedema, elevated blood pressure and exacerbation of heart failure. Short-term uses of NSAIDs are, however, well tolerated if the patient is well hydrated and has good renal function with no heart failure, diabetes or hypertension (2).

Erythromycin, especially when prescribed to patients with CKD, has been associated with reversible hearing loss. Patient with CKD receiving doxycycline and azithromycin require no changes to dosage. Tetracyclines can increase nitrogen retention and acidosis and are best avoided. Benzylpenicillin has significant potassium content and may be neurotoxic. Penicillins (other than phenoxymethylpenicillin and flucloxacillin) and metronidazole should be given in lower doses, as high serum levels can be toxic to the central nervous system (2). Other potential medication complications are listed in ***table 3*** (6).

*Effect of Haemodialysis:*

Patients undergoing haemodialysis may experience difficulties in chewing, swallowing, tasting and speaking. There is an increased risk of oral disease including lesions of the mucosa, gingiva and tongue. Bacterial and fungal infections such as candidiasis, dental caries and periodontal disease may occur. Interdialytic weight gain occurs due to increased fluid intake and there is an overall reduction in the quality of life. Unfortunately there is no effective treatment for hyposalivation in patients on chronic haemodialysis. Uraemic stomatitis is now a rare complication of CKD (2).

*Use of Anesthesia and Sedation:*

Local anaesthesia (LA) is safe unless there is a severe bleeding tendency. For conscious sedation, relative analgesia is preferred. If IV sedation is necessary, veins that are at or above the elbow should be used due to the risk of consequent fistula infection or thrombophlebitis. A cannula should never be placed into the arteriovenous fistula arm. Midazolam is preferable to diazepam due to lower risk of thrombophlebitis (2).

Anaemia is frequently found in CKD patients as a result of toxic suppression of the bone marrow, a lack of renal production of erythropoietin and/or iron deficiency from blood loss in the gut. General anesthesia is contra-indicated in patients with hemoglobin levels below 10g/dL.

*Growth retardation:*

Growth retardation is a significant characteristic in children with chronic renal failure. It has been found that the earlier the onset of chronic renal failure, the more severe the growth delays. There

are many factors that cause growth retardation including reduced food intake and chronic metabolic acidosis. In order to compensate for their reduced calorie intake, these children are generally encouraged to increase their intake of carbohydrates mainly in the form of refined carbohydrates, sweets, soft drinks and other sweetened food products. Hence the clinician must closely review these patients, as even though patients with CKD have low caries risk due to greater salivary protective factors, once these patients are transplanted and renal function becomes normal, they will be at an increased risk of caries, especially if the high-cariogenic diet continues in addition to the presence of enamel hypoplasia and poor oral hygiene (23).

## Conclusion

The prevalence of kidney disease in the paediatric population is increasing worldwide. Oral and systemic complications can occur due to the underlying disease or its treatment. Dental management is complicated as a result of these systemic consequences however with the use of appropriate precautions, dental management can be effectively and safely provided to these medically compromised patients.

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# Federal President's Report

*Tim Johnston*

**2017 has hit with a bang, I don't know what happened to the break over the Christmas period, that went in a blink and we are all back at it probably still needing a bit more of R&R.**

The East coast of Australia is sweltering with record heat waves, the West is experiencing record rain and according to my weather app, New Zealand is perfect as usual.

The RK Hall meeting is just around the corner, we are looking forward to hearing Dr Ari Kupietzky, current Secretary General of the International Association of Paediatric Dentistry and a leader in paediatric biomimetic dentistry. Ari will be supported by local speakers covering a great and varied range of topics. Of course the social program is equally important (to me) and the Society as a whole looks forward to enjoying an excellent scientific program and a wonderful catch up with old and new friends. I certainly encourage all members to make their way to Auckland in March.

It is a pleasure to be able to acknowledge and congratulate the 2016 winners of the Louise Brearley Messer Postgraduate and Undergraduate Essays, Dr Maansi Juneja of the University of Sydney and Miss Joon Soo Park of the

University of Western Australia respectively. The Essay questions are set by the ANZSPD Federal councilors and quite a lot of time is spent discussing and choosing the question for each. The questions are challenging, often left of field and all entrants give the examiners a difficult task to select the final winner. Congratulations to all those who submitted an essay and congratulations to Maansi and Joon Soo, I look forward to reading your essays in future editions of Synopses.

ANZSPD has remained active in providing submissions to different dental and government bodies through the second half of 2016 and into 2017. ANZSPD responded to the ADA's request to lobby the Australian Federal Health Minister to reconsider the dropping of the Child Dental Benefits Scheme. Just this week the Turnbull Government has backed up their earlier decision to retain the CDBS and reinstated the $1000 cap. Whilst this is a win for the dental professional and the child community, we are still no further ahead addressing the 0-4 age group and the children that require treatment under general anaethesia. I also wonder how the decision will affect the planned Child and Adult Public Dental Scheme announced during the last election campaign and worry the re-instated CDBS cap will reduce funding where it is equally needed. ANZSPD will move back to the old crusade of trying to address the unseen children, two steps forward, two steps back.

The combined ANZSPD AAPD website has had a complete overhaul and has finally become what John Winters sent out to build a number of years ago. It has been said before and I will do it again, a huge thanks needs to be extended to John for his persistence, patience and maybe a fair degree of bloody mindedness to produce what we have now. John is close to handing over the running of the website to the Website committee, a bunch of young tech savvy colleagues who will take a great facility and make it even better.

I urge all those who have not renewed their membership to do so through the website. It is easy and fast. I especially urge our New Zealand members to do the same. The problems of the past have been sorted out and your renewal is now as straight forward as your Australian colleagues.

ANZSPD was invited to submit an article outlining the history of ANZSPD and the current state of children's oral health in Australia and New Zealand. Thank you to Drs Peter Gregory and Carmel Lloyd in their assistance with the article in providing the needed information. Part of the article was an 'interview with the President', the last question being "What are the Association's future projects ?" I answered survival. With all the complexities that go along with it. The readers of this report are likeminded and see paediatric dentistry as a very important part of the dental profession, if not the most. But the profession as a whole I don't think see it this way. Sadly this is reflected by the Australian Dental Association 37th Federal Congress in Melbourne in May 2017. The scientific program is extensive and interesting, however there is zero paediatric dental content. I think we can admit paediatric dentistry is not the glamour area of our profession, although we all marvel at a nicely placed stainless steel crown, others see more beauty in an implant supported anterior ceramic restoration. Some of these being provided today are so 'real' that I see their reason. With the economic pressures dentists face today, they need to be very careful where they spend their money for continuing education and will and should focus on treatments that they can provide well to their patients and obviously provide the practitioner with optimal financial return. It is pleasing that ANZSPD continues to grow as a society, in membership, standing within our own and the international community and continues to attract the highest caliber of international and local speakers to our meetings. The RK Hall meeting in Auckland is a beautiful example of this. And despite maybe discussing the merits of diammine silver fluoride rather than higher income implant dentistry, ANZSPD's projects, from our biennial meetings right through to local evening meetings listening to postgraduate presentations, the Australian and New Zealand Society of Paediatric Dentistry is not only surviving, we are flourishing.

Thanks as always to the hard working Councilors, Federal Secretary and Federal Treasurer and to all branch executives. In 2017, the Future for ANZSPD looks bright.

I wish every one a safe year and see you in Aotearoa.

## Branch Report New South Wales

ANZSPD NSW Branch had an excellent year of continuing education and professional development in 2016. We held three very successful dinner meetings in 2016. Dr Bernard Koong lectured on the use of "Diagnostic imaging in Paediatric Dentistry. The importance of cone-beam CT was discussed in detail. Dr. Peter Wong spoke at the second meeting of the year on medico legal issues. Peter's topic was "Negligence: what you weren't taught at the university" and spoke in depth about medico-legal issues and how evidence based practice is very important. This lecture also re-enforced that communication is a key and integral part of provision of dental care to our patients.

The third and the last dinner lecture for year 2016 was delivered by our current ANZSPD Federal President Dr Timothy Johnston, Tim's topic "Are stainless steel crowns still the gold standard?" generated considerable amount of discussion. This lecture not only discussed about stainless steel crowns but various aspects of Paediatric Dentistry with a special interest in diagnosis and restorative care using loupe and microscope magnification.

We have planned three meetings for the year 2017 including the AGM. The venue for ANZSPD lecture meetings for 2017 will The Mantra Hotel, 10 Brown Street, Chatswood NSW 2067.

The lecture meetings for Year 2017 have been finalised and will be held on 14th March, 6th June and 17th October(AGM). Our speakers include Dr Michael Brosnan from University of Otago, Dr Harleen Kumar, Head of Paediatric Dentistry, Sydney Dental Hospital an Dr Chetan Pandit, Head of Respiratory Medicine at Westmead Children's Hospital.

Under the leadership of our current branch President, Dr Naveen Loganathan, we look forward to a good year ahead. Dr Diane Tay who is our new treasurer has been working hard to keep our finances in order. As always, Dr Soni Stephen who is the NSW Federal Councillor has kept us up to date with all Federal ANZSPD issues.

We look forward to meeting you all in Auckland in March 2017. Have a great year.

*Dr Prashanth Kumar Dhanpal*

## Branch Report: Western Australia

The ANZSPD WA Branch would like to wish everyone a happy start to 2017. There have been a couple of changes to the paediatric family in WA. Dr Joy Huang, a recent graduate from the University of Melbourne has returned to Perth and taken up the reigns from Dr Susan Wong to be our full time Senior Dental Registrar at Princess Margaret Hospital. A big thank you to Susan for her dedication, hard work and mentoring during her time as the Senior Dental Registrar at PMH. Susan has now transitioned into Private Practice working for Dr Marilyn Lobo.

More good news from Western Australia, is that Dr Chat Neboda has been awarded the Alistair Devlin Memorial Scholarship. This scholarship is provided to encourage and assist a student in their final year of the degree of Doctor of Clinical Dentistry. Chat has taken this opportunity to attend Our Lady's Hospital for Sick Children in Dublin, Ireland and Boston Children's Hospital in the United States of America. At the end of her externship, she will be presenting her research at the International Association of Dental Research Conference in San Francisco in March 2017.

The West Australian Branch has a terrific program lined up for 2017 and would encourage all members to visit sunny Perth to attend our meetings. Our meetings are published on the ANZSPD website, and ticket registration is through anzspdwa.eventbrite.com.au.

Our first meeting of the year is always a social event with partners. The evening will be set in the relaxed ambiance of Manuka Woodfire Kitchen where you will be treated to a high quality meal showcasing local produce. We are proud to announce that the Louise Brearley Messer Undergraduate Essay Competition was awarded to Joon Soo, from the University of Western Australia. Joon will be awarded her Prize by Dr Tim Johnston at this event. Later on in the evening our guest speaker, Mr Mike Wood will be giving a presentation on his experiences preparing and climbing the Himalaya including the Everest region.

The postgraduate presentation evening will be held on the 25 May at an inner city foodie haunt. Our paediatric postgraduate students provide 10 minute presentations on either their research or interesting case presentations. This evening is well attended, show casing a wide range of topics.

On the 21st-22nd July, our mid-winter meeting will be held at Pullman Bunker Bay Resort in Dunsborough, located 3 hours South of Perth. Dunsborough is considered a favourite "down south" family holiday destination, renowned for its stunning coastline and award-winning wineries and breweries. The theme of the Scientific Meeting this year is the Orthodontic-Prosthodontic Management of children with Hypodontia, Oligodontia and Anodontia. As with previous meetings, these lectures will be interspersed with a potpourri of short 15-20 minute presentations by various members of our group.

This year's Scientific Meeting will be held on the 15th September. We have been fortunate to have Professor Lars Andersson accept our invitation to be our Keynote Speaker on the topic of "Managing the Complications following Traumatic Dental Injuries". Further details will become available shortly on the Eventbrite system. He will also provide an afternoon seminar, to our Postgraduate Students on the 14th September.

We would like to wish all members a great start to the new year. For any questions regarding our program, please do not hesitate to contact our ANZSPD WA Branch Secretary, Dr Rebecca Williams at anzspdwa@gmail.com

*Vanessa William*

## Branch Report South Australia

2017 looks to be another thought-provoking year for the ANZSPD SA Branch! We have four dinner meetings planned covering a range of topics. Our first meeting will cover anxiety in young children, lead by the Head of the psychiatric inpatient ward at the Women's and Children's Hospital, Dr Hsu-en Lee. In May, we look forward to a presentation by Professor Kaye Roberts-Thomson on "Fluoride - friend or foe?". Our third meeting of the year, titled "All in a night's work - tonsils, adenoids, snoring and sleep apnoea in children", will cover a topical area of dentistry. For our final meeting of the year, radiographic presentation of oral and maxillfacial cysts and tumours will be revisited.

Our dinner meetings this year will also be interspersed by a September seminar at the Adelaide Zoo. Our keynote speaker is Professor Lars Andersson, who will be presenting in both South Australia and Western Australia. Professor Andersson is synonymous with dental traumatology and will cover dental trauma in the permanent dentition comprehensively. It will also be very exciting listening to Dr James Lucas as well as other local speakers discuss aesthetic crowns, frenectomies and more.

*Gwendolyn Huang*

# Major aphthous ulceration in early onset inflammatory bowel disease: A case report

*Dr. Venkatesh Bhardwaj (Registrar, Department of Paediatric Dentistry & Orthodontics, Westmead Centre for Oral Health, Westmead, NSW)*

## Introduction

Inflammatory bowel disease (IBD), encompasses Crohn disease (CD) and ulcerative colitis (UC) and is typified by chronic inflammation of the gastrointestinal tract in children and adults. Both these spectrum of diseases are thought to arise in individuals that are genetically predisposed with disruption of epithelial and mucosal barriers, likely in the context of aberrant inflammatory signals, loss of tolerance and environmental triggers[1]. Although precise population based data on IBD is lacking, the incidence peaks in the second decade of life[2]; among all patients with IBD, 20%-30% with CD and UC are diagnosed in the paediatric age group[2,3]. Diagnosis of IBD in infancy is rare, with little information available regarding presentation below two years of age. In patients with IBD, extra-intestinal complications including oral manifestations occur in 25% of cases, with the oral mucosa affected more so in CD in 0.5% to 20% of cases[4]. Oral manifestations are often considered an extension of the granulomatous disruption into the oral cavity, clinical presentation often including swelling of labial tissue, and cobblestone appearance of the gingiva and mucosa. Though oral aphthous ulcers are frequently reported to occur in CD and UC, in the general population they are known to be the most common cause of oral ulceration in adults and children; and reported to affect up to 40% of children[5]. Precisely measuring the impact that IBD has, in the aetiopathogenesis of oral aphthae is a challenge as other causes such as nutritional deficiencies in iron, folic acid and vitamin B12 have also been reported to cause recurrent aphthous stomatitis[6]. As the incidence of IBD increases in the paediatric population, it is possible that the number of presentations with oral manifestations will also rise. Knowledge of the disease, will therefore help the clinician to recognise patients and refer them for appropriate specialised care. The following is a case report of a child that presented with symptoms of major oral aphthous ulceration, decreased oral intake and failure to thrive.

## Case report

A 17 month old boy was brought by his parents to the Emergency department of the Children's Hospital Westmead (CHW), Sydney, NSW with a two week history of oral ulceration and a week of reduced oral intake and concurrent diarrhoea. A background of 2 months of intermittent fevers, treated with multiple courses of antibiotics for suspected upper respiratory tract infections and otitis media was reported, along with a predisposition to atopy with eggs and nuts and history of food protein-induced enterocolitis syndrome (FPIES). The boy was a first child, born full-term via caesarean section, to non-consanguineous parents both with a history of atopy (rhinitis and eczema), but no family history of any immunodeficiencies, inflammatory bowel disease or oral ulceration. There was no reported recent contact with sick individuals. Given the complex history and presentation, multidisciplinary input involving dental and medical teams was sought. On examination, the child was alert but unwell, febrile and miserable. Oral examination revealed two large ulcers on the lower lip and tip of the tongue *(Figs. 1,2)*, each greater than one centimetre in diameter. A generalised eczematous rash was noted with no genital or perianal involvement. Initial investigations included a full blood count which were mostly unremarkable (WCC, neutrophils, LFT, vitamin B12, folate, viral swab)[a] except for a microcytic & hypochromic haemoglobin level and slightly raised ESR and CRP[b]. Additionally stool samples were taken to rule out infective causes. In the days following initial presentation, the patient continued to get unwell and avoid oral feeds, necessitating insertion of a nasogastric tube to continue feeds. Pain was controlled with MS Contin (2g bds) and oxycodone (prn – max 6 doses daily) by the pain management team. Additionally, due to the persistence fevers and possibility of underlying bacterial causes, IV vancomycin and cefotaxime was commenced. Due to progressive worsening and avoidance of oral feeds, endoscopy of the gastrointestinal tract and punch biopsy of the oral ulcerations, under general anaesthesia was performed which revealed, several, giant aphthous ulcerations *(Fig 4)* of the gastrointestinal mucosa, suggestive of an early onset inflammatory bowel disorder (Crohn or Crohn like disease). Viral serology and bacterial cultures were both negative. In liasion with the gastroenterology team and dietetics, solid intake was ceased and the child maintained on breast feeds and Neocate®[c], along with adequate pain management. The child's condition improved significantly following this with complete recovery and oral ulcers eventually healed well without any therapeutic intervention being required, with further testing for genetic and immunological markers (IL-10) as well as enzymes (TPMT[d]) currently under way, in order to determine long term prognosis and institute a management plan involving therapeutic immunomodulatory drugs.

## Discussion

Ulceration of the oral mucosa is common and said to occur in up to 40% of the paediatric population[5]. Though majority of cases of oral ulceration are said to be idiopathic, they can be associated with underlying systemic inflammatory or autoimmune conditions that may have genetic and/or environmental influences, as was seen in this case. In young children, such as in the case of this 17 month old child, quality of life may be affected to the point where there is failure to thrive due to discomfort and reduced oral intake[7]. In a child patient, oral ulceration may be caused due to a number of reasons and it is important to be able to approach the likely causes in a methodical manner.

Recurrent aphthous stomatitis (RAS) remains the most common ulcerative disease of the oral mucosa, often presenting as round, painful, shallow ulcers with well-defined erythematous margins and yellow-gray pseudomembranous centre[5]. Other diseases that cause oral ulceration that may prove hard to differentiate from RAS include Behcet syndrome, cyclic neutropenia, intraoral herpes infections, HIV-related ulcers or inflammatory bowel disease. It is nonetheless, important to distinguish RAS from ulcers caused due to an underlying systemic disorder.

In children between ages of 6 months to

a WCC – White cell count - Live function tests
b CRP – C reactive protein; ESR - erythrocyte sedimentation rate
c Infant formula commonly used in children with allergy
d Thiopreene methyl transferase is an enzyme, that plyas a crucial role in metabolism of thioprene drugs, and is encoded by the TPMP gene Testing for this gene is commonly recommended prior to commencing immunomudulatory drugs such as azathioprene

five years, presentation with non-specific systemic symptoms such as malaise, fever and lymphadenopathy succeeded by oral ulceration is often suggestive of a primary oral herpes simplex virus (HSV) infection that commonly occurs early in childhood. Viral serology, when performed will test positive to HSV DNA, although these tests tend to be reserved for evaluating immunocompromised patients with atypical lesions for definitive diagnosis8. In this case, swabs from the ulcer base tested negative for HSV DNA. When systemic features accompany atypical oral ulceration, the priority is to exclude serious underlying disease, as was the case in this child with a background history of intermittent fevers and accompanying symptoms of diarrhoea, thereby warranting further investigation, using a multi-disciplinary approach.

A full blood count is commonly the first screening test performed in order to look for markers of disease such as WCC, CRP, ESR, LFT's which were unremarkable in case of this child. Cyclic neutropenia, a rare disorder that may present in childhood has also been associated with oral ulceration in periods with neutropenia. Given the normal neutrophil count during this presentation, this was able to be ruled out in our patient along with some of the periodic fever syndromes such as PFAPA[e], that also tend to show neutropenic states.

Beyond the full blood count, haematinics may be employed to rule out nutritional deficiencies (iron, ferritin, folate, B12, Zn and Mg). The microcytic, hypochromic film in this child was typical of an iron deficiency anaemia. Though the role of nutritional deficiencies (low serum levels of iron, folate, zinc or the vitamins B1, B2, B6 and B12 ) as a cause of oral aphthous ulceration has been suggested6, it has yet to be shown conclusively, and more so in children. It is worthwhile noting that the accompanying prodrome and symptoms of diarrhoea as were seen in this child are not commonly associated with ulceration due to nutritional deficiencies. It is a possibility that nutritional deficiencies in this child may have been caused secondary to underlying inflammatory enteropathies, rather than as sole aetiological factors of the oral ulceration. That the oral ulcers did not abate with iron supplementation may also be indicative of the underlying aetiology not being predominantly nutritional.

The most notable medical disorder associated with oral aphthous ulceration is Behcet syndrome typified by recurring oral, genital and eye lesions[9]. Although predominantly a disease seen in adults, several cases have been reported in children[10]. Other variants of Behcet syndrome such as relapsing polychondritis of MAGIC[f] syndrome have been outlined in the literature however the age at presentation and accompanying symptoms generally ruled out these disorders in our case.

Patients with IBD often suffer from aphthous ulceration of the oral cavity. However, since these lesions also occur commonly in the general population, they are considered non-specific with regard to IBD. In a retrospective study, a high prevalence of disease-specific oral lesions was found in children with CD[11]. Additionally, this study also found granulomatous lesions were noted in oral biopsy specimens in over 75% of cases suggesting the significance of the oral cavity in obtaining diagnostic tissue in children with symptoms of IBD[11]. Despite such findings, and mainly due to a dearth of adequate published evidence, oral ulcers in the absence of intestinal manifestations are rarely subjected to biopsy. However in this case, the onset, age of the patient, atypical ulceration and failure to thrive were sufficient for the medical team to justify endoscopic biopsy of the lesions under general anaesthesia, in order to confirm diagnosis. The presence of ulcers throughout the GI tract seen during endoscopy along with biopsy findings in this case were highly consistent with an inflammatory condition such as Crohn disease, however it remained unclear if an underlying immunological abnormality was also present. Current research in the field of early onset IBD revolves around investigating mutational defects in the expression of immunological proteins, with infants that show impaired IL-10 signalling frequently presenting at less than 3 months of age with a severe and progressive disease[1]. Our patient's symptoms resolved within a week following biopsy after which solids were ceased and an infant formula (Neocate®) for children with allergies was instituted. Adequate weight gain was noted at discharge 3 weeks following initial hospital admission, with monthly follow up in order to complete immunologic and genetic testing organised.

The importance of early diagnosis of inflammatory conditions in children is that it allows adequate nutrition for optimal growth and development. Untreated disease therefore, can have a significant knock on effect delaying puberty, which may result in permanent growth impairment. Inducing disease remission before the onset of puberty and maintenance of remission during the pubertal years, is known to be crucial to avoid unacceptably short stature due to a missed pubertal growth spurt[12]. Early identification, as was in the case of this child allows for optimal management, which may afford a chance at achieving remission.

Medical management of IBD in the paediatric patient is supported by use of anti-inflammatory drugs including steroids and non-steroidal anti-inflammatory drugs similar to those based on 5-aminosalicylic acid (5-ASA)[g]. In severe cases, this may involve a combination of steroids such as prednisolone and immunomodulators such as azothioprene[13]. The challenge with the use of steroids in children lies in its interference of normal growth and development. Newer "steroid sparing biological agents" that target the immune machinery have been trialled successfully and have benefits in their use in the growing child.

In the case presented here, due to the resolution of the condition following introduction of non-allergy infant formula, management of pain was the only therapy required. Despite the phenotypical presentation of Crohn disease in this child, the possibility of an underlying genetic cause may mean the inability to achieve a precise diagnosis immediately. Recent research has suggested the role of an anti-inflammatory cytokine IL-10 in maintaining tolerance against possible pathogenic invaders1. As a consequence, tests for IL-10 genotype and TPMT enzyme are currently underway.

The tendency for GIT bleeding in these patients at a chronic low level may cause a higher chance for iron deficiency anaemia which may contribute to oral ulceration, angular chelitis, candidal infections and glossydynia[13]. Additionally, therapeutic management of the condition with steroids and other immunosuppressives can lead to increase in opportunistic species. The importance of maintaining good oral hygiene and regular professional dental care must be reinforced with patients and parents alike. It is important to realise that these patients often need to maintain calorific needs involving frequent oral intake and care must be taken to not dispense contradictory advice, rather collaborate with the dietitian in order to institute early caries prevention. In older children, symptomatic care may

e Periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis also known as Marshall syndrome
f Mouth and genital ulcers with inflamed cartilage
g Mesasalazine

involve rinsing with chlorhexidine (0.2%) or benzydamine hydrochloride (0.15%) to manage pain associated with oral ulceration.

Though IBD in infancy is rare, the first presentation may be to the dentist with recurrent oral ulceration. The paediatric dentist may therefore be in a position to recognise features of the condition and highlight the possibility of a systemic disorder thereby facilitating timely referral to a paediatrician to enable early identification and management.

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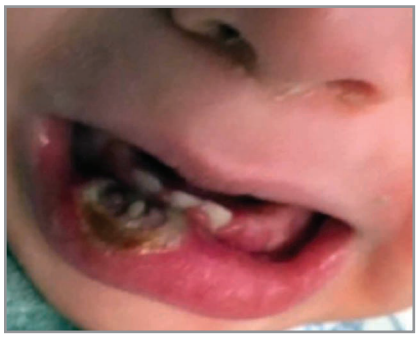
## List of figures

*Figs 1, 2: At initial presentation – Ulceration on lower lip and tip of tongue*



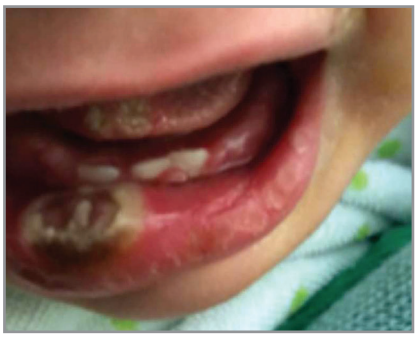


*Fig. 3: Ulceration on tongue & lip prior to biopsy*

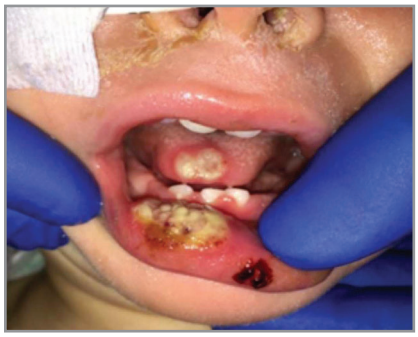
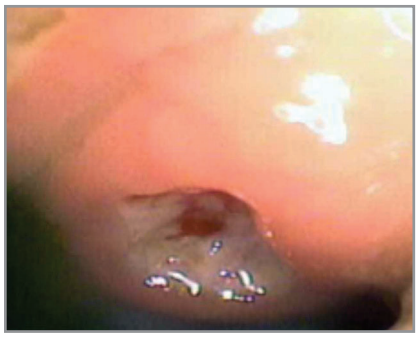
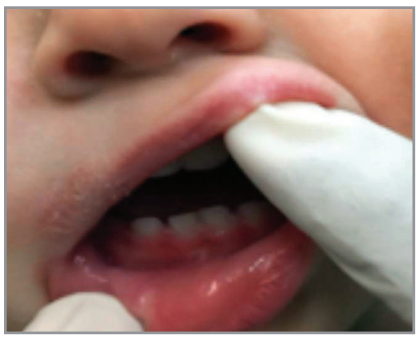
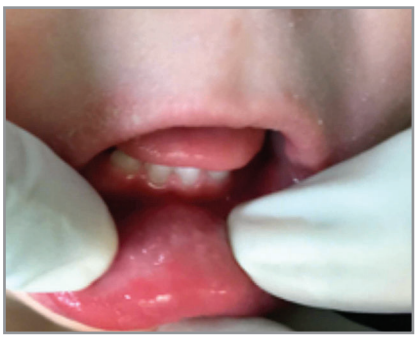


*Fig. 4: Endoscopic image – Giant aphthous ulcer in colon*



*Figs 5, 6: Complete resolution of lip and tongue at 4 month follow up*





# Oral health related quality of life in Australian children with orofacial cleft and their families

*Dr Caitlin Agnew*

## Introduction

Children affected by orofacial clefts may experience compromised health, psychosocial well-being and quality of life[1]. Fortunately, there has been a move towards a socio-environmental model of healthcare which has been complemented by growing interest in the assessment of OHRQoL. However, despite the core OHRQoL issues in cleft children, there remains limited information regarding their OHRQoL and further research is called for[2]. In this modern era, it is important that children are involved in research efforts, with research performed with them and not on them, highlighting the importance of gaining their perspectives and experiences[3]. This article will give a brief overview of OHRQoL in children with cleft as well as details of a clinical research study which is currently being conducted in Sydney titled "Oral health related quality of life in Australian children with orofacial cleft and their families."

## Orofacial clefts – overview and management

Orofacial clefts are among the most common visible birth defects worldwide[4]. Due to their distinct developmental origins, OFC are categorized as cleft lip with or without cleft palate (CL/P) and isolated cleft palate (CP)[5]. In Australia, the incidence of CL/P is 1.2 per 1000 births and for CP is 1.4 per 1000 births[6].

Children with cleft experience prolonged and extensive treatment and rehabilitation, which begins at birth, and continues into the adult years. Treatment begins in the first year of life, and is comprised of primary surgical interventions such as closure of the lip and palate. This is followed by a myriad of assessments carried out by an interdisciplinary team to address certain characteristics such as facial appearance, speech and the dentition[2]. Secondary surgical treatment, such as alveolar bone grafting, orthognathic surgery and rhinoplasty may be indicated in the early to late adolescent years[7]. Not without risk, multiple surgeries are carried out under general anaesthesia before these children reach adulthood[8]. The final aim of treatment is to achieve closure of the cleft, favourable facial growth, suitable aesthetics and function, as well as the development of speech and hearing to enable good communication.

## Oral Health Related Quality of Life (OHRQoL)

Fortunately, there has been a shift from the purely surgical treatment of patients with cleft, to a more holistic, socio-environmental approach[9]. The health profession in the last decade has seen the increasing use of the term 'oral health related quality of life' and in this modern era, it is imperative that traditional measures of health be supplemented by QoL measures, obtained from patients, that captures their experiences and concerns[10,11]. Today, QoL outcome measures are recognized as valid parameters in patient assessment in nearly every area of healthcare, including oral health[12].

Oral Health Related Quality of Life (OHRQoL) is an important aspect of QoL and is a report given by patients to assess how oral disease affects daily functioning and psychosocial wellbeing [13,14]. OHRQoL is a multidimensional concept, and is increasingly recognized as an integral part of health and well-being[15]. More recently, OHRQoL has been identified as a key health priority, highlighting the profound impact that oral health may have on a child's QoL[16]. The common dimensions studied in OHRQoL instruments, as well as examples of items associated with each dimension are shown below.

## OHRQoL in children with cleft

Until recently, impaired QoL as a consequence of oral conditions had received minimal attention. Many children with cleft are significantly burdened by compromised oral health, including an increased prevalence of dental anomalies, dental crowding, malpositioned teeth, skeletal discrepancies, dental caries and periodontal disease[17-19]. A recent systematic review by Antonarakis and colleagues reported that children with cleft, have poorer OHRQoL compared to the general non-cleft population[20]. Furthermore, the literature has highlighted that the dimension of OHRQoL most impacted in these children is functional wellbeing[11]. Poorer functional well-being has been reported to be due to issues with eating, being understood, and keeping their teeth clean[13]. These children are also prone to compromised emotional well-being[13,21] and school-environment well-being[2,22], due to aesthetic concerns, impaired communication skills and teasing by peers.

## Oral health related quality of life in Australian children with orofacial cleft and their families

The aim of this research is to investigate the OHRQoL in Australian children with orofacial cleft, and to analyse whether a child's oral condition influences family functioning. This is the first study of its kind conducted in an Australian cohort of children, and was carried out from September 2015 to October 2016 at the Cleft Clinic at The Children's Hospital, Westmead and at the Paediatric dentistry and Orthodontic department, Westmead Centre for Oral Health. 223 children aged 7-18 years with orofacial cleft completed a Child Oral Health Impact Profile (COHIP) questionnaire and 223 caregivers completed a COHIP questionnaire (regarding their child), and a Family Impact Scale (FIS) questionnaire.

## The Child Oral Health Impact Profile (COHIP)

The COHIP is an established OHRQoL questionnaire used to assess OHRQoL in children aged 7-18 years with varying oral conditions[23,24]. It has been shown to

*Figure 1: Dimensions comprising oral health – related quality of life[15]*

have excellent scale, test-retest reliability and both discriminant and concurrent validity[23], therefore representing a useful psychometric tool for measuring OHRQoL [25]. It comprises a questionnaire for the child to complete and one for the parent, and has been translated into multiple languages, making it applicable to different populations worldwide[26,27].

The major limitation of the COHIP is its length. The COHIP short form (COHIP-SF) is a considerably shorter questionnaire which reduces respondent burden and allows for easier, more efficient application in clinical practice and research[24]. The COHIP-SF includes 19 items, measuring three domains, namely; Oral Health (five items), Functional Well-Being (four items) and Socio-Emotional Well-Being (ten items). The COHIP-SF has been found to have good psychometric properties, which are comparable to the original COHIP[24]. For further information regarding the COHIP-SF please refer to the article: Reliability and validity testing for the Child Oral Health Impact Profile-Reduced (COHIP-SF)[24].

## The Family Impact Scale

The family impact scale (FIS) was developed and evaluated by Locker and colleagues 28 and assesses the impact of how having a child with a cleft, influences parental and family activities, parental emotions, conflict within the family and family finances[29]. The short-form FIS (FIS-SF) was created by shortening the FIS from 14 to 8 items, and it has been found to have acceptable concurrent validity, responsiveness, reliability and validity[30].

For further information on the FIS-SF please refer to the article: Short-form versions of the Parental-Caregivers Perceptions Questionnaire and the Family Impact Scale[30].

## Research Objectives

- To determine overall OHRQoL in Australian children with cleft
- To determine oral health, functional and socio-emotional well-being, using the COHIP-SF measure
- To determine parental perceptions of children's OHRQoL and how it compares to their child's reported OHRQoL
- To assess if there is any influence of age, gender, cleft type, private health insurance, number of past cleft-related surgeries or rural living location on OHRQoL
- To determine whether a child's oral condition influences family functioning using, the FIS-SF measure

## Analysis of results

For each COHIP question, subjects are asked how frequently they have faced a positive or negative experience relating to their teeth, mouth or face during the past 3 months. Responses are made on a Likert-type scale (0 = never, 1 = almost never, 2 = sometimes, 3 = fairly often, and 4 = almost all of the time). The COHIP allows for the calculation of each subscale, as well as an overall COHIP score which ranges from 0 (worst OHRQoL) to 76 (best OHRQoL)[31].

For each FIS question, parents are asked to relate each question to the last three months on a Likert scale which is scored from 0–4. An example of a question in the FIS-SF is: during the last 3 months, because of your child's teeth, lips, mouth or jaws, how often have you or another family member, felt guilty or had disrupted sleep?

The collected clinical and demographic data will be described using standard statistical methods.

## Conclusion

Incorporating OHRQoL into clinical practice has essential benefits for patients and their families [15]. Measuring OHRQoL enables an understanding of treatment needs and how oral issues impact QoL. OHRQoL instruments have a multitude of substantive applications for the field of paediatric dentistry and orthodontics and should be considered in future research efforts so that understanding of the relationship between oral health and general health and wellbeing is enhanced. It is also important in the modern era, that research is conducted with children, and not on children, highlighting the importance of gaining their perspectives and experiences of oral health.

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# 'My baby's loosing teeth!' An interesting presentation of Hypophosphatasia in a toddler

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## Introduction

Hypophosphatasia (HP) [1] (OMIM 146300, 241500, 241510) is a rare, inherited disorder characterized by defective bone mineralization and a deficiency of tissue-nonspecific alkaline phosphatase activity (TNAP)[2]. The mode of inheritance may be either autosomal dominant or recessive [3]. There are six recognised forms of HP ***(Table 1)*** despite a continuum of severity reported in the literature which ranges from very mild to severe, which is incompatible with life [3, 4]. The reported prevalence for Perinatal HP (severe) is 1:100 000 with milder forms considered to be much higher. This is likely given cases that exist undiagnosed, due to very mild symptoms.

## Clinical manifestations:

As HP exists across a spectrum, clinical expression can be very mild affecting teeth only to more obvious symptoms such as delay in walking, short stature, skeletal deformities, bone pain and pathological fractures. In the perinatal form, stillbirth occurs due to failure of bone mineralization [10]. Generally, the earlier the presentation the more severe the disease. While some symptoms are more pertinent to one subtype it is recommended that all patients be counselled appropriately due to potential long-term consequences. A number of dental findings in both the primary and permanent dentitions have been reported in the literature. Often the first clinical manifestation involves the primary teeth, which consequently results in dentists/dental specialists being the first health care professionals to assess early clinical signs in these patients.

## Case report

A fit and healthy girl (SK) eighteen months of age was referred from a general dental practitioner (GDP) to the Specialist Paediatric and Orthodontic dental department for assessment. SK's parents were concerned about the loss of her lower anterior teeth, which had exfoliated at 15 months of age (3 months prior). SK's medical history included a complicated pregnancy due to maternal hypertension resulting in early gestational age of 35 weeks. SK was the only child and no family history of dental abnormalities was given. SK's mother presented both the exfoliated teeth at consultation, and the entire crown and root form for each tooth was evident ***(Figure 1)***. There was no traumatic dental injury (TDI) history reported. According to her mother, the lower primary central incisors (71, 81) had erupted at 7 months of age and the upper primary central incisors (51 and 61) had erupted at 10 months of age.

Clinical examination revealed the patient was dentally age appropriate with healthy soft tissues and a caries-free primary dentition. Of note, missing teeth included 71 (lower left primary central incisor) and 81 (lower right primary central incisor) and Grade I mobility was associated with the upper primary incisor teeth (51 and 61). There were no abnormal findings of the erupting teeth (52,62,72,82,54,64,74,84) and generally, oral hygiene was optimal, (***Figure 2)***.

*Table 1 [9]*

| *Subtype* | *Inheritance* | *Clinical Manifestations* |
|---|---|---|
| Perinatal (lethal) (OMIM #241500) | Autosomal recessive | Marked impaired bone mineralisation *in utero*, blue sclerae, skin-covered osteochondral projections affecting limbs (often diagnosed)[4] respiratory complications. Often lethal. |
| Prenatal (benign) | Autosomal recessive or autosomal dominant | Limb deformities (may improve during 3rd trimester of pregnancy). Spontaneous improvement of skeletal defects reported[5,6]. |
| Infantile (OMIM #241500) | Autosomal recessive | Onset before 6 months of age. Respiratory complications, premature craniosynostosis, demineralisation, rachitic changes in the metaphyses, hypercalcaemia[1], short statue, premature loss of primary teeth. Mortality is often related to respiratory complications. |
| Childhood (OMIM #241510) | Autosomal recessive or autosomal dominant | Onset after 6 months. Skeletal deformaties and fractures, short statue, premature loss of primary teeth. Often self-limiting but may re-appear in adulthood[1]. |
| Adult (OMIM #146300) | Autosomal recessive or autosomal dominant | Presents in middle age often with stress fractures, thigh pain, chondrocalcinosis and osteoarthropathy. Patients may reportr premature loss of primary +/or permanent teeth[7,8]. |
| Odontohypophosphatasia (OMIM #146300) | Autosomal recessive or autosomal dominant | Premature exfoliation of primary teeth (most commonly the incisors). Often no skeletal manifestations. |

*Figure 1: Exfoliated teeth*

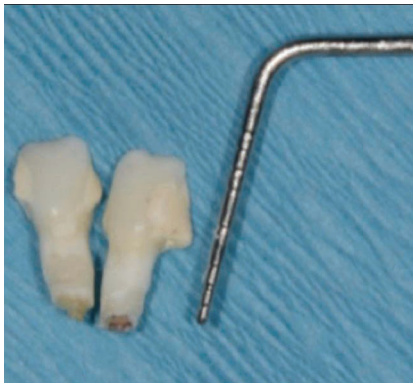


*Figure 2: Clinical photographs at consultation*

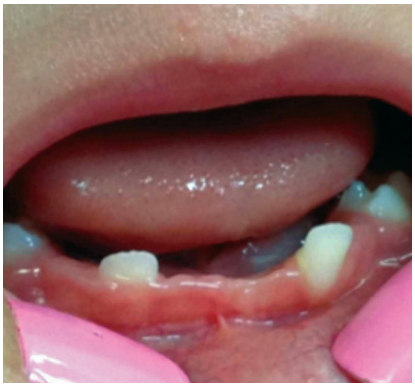
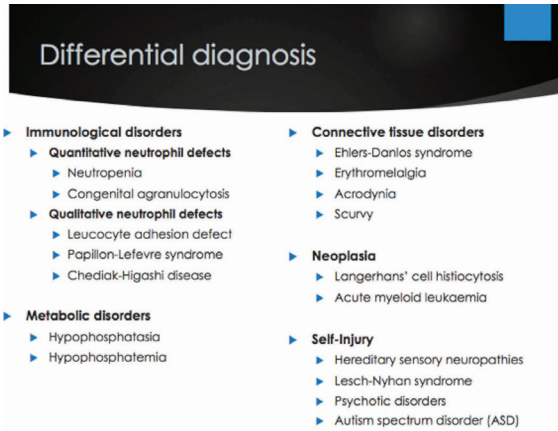


*Figure 3: Diagnostive Sieve*



A full blood count analysis was within the normal ranges, however very low levels of alkaline phosphatase (ALP) 59 U/L (normal: 120-350) and high serum phosphate levels 2.28 (normal: 1.0-2.0) were detected.

## Differential diagnosis:

There are a number of conditions and disorders causing early exfoliation of primary teeth. Both local and systemic factors can contribute to this finding, including serious and potentially life threatening conditions and thus a thorough investigation is warranted. The following list of differential diagnoses was considered in this case as follows ***(figure 3)***.

Based on the clinical findings and the lack of systemic involvement for SK a provisional diagnosis of Hypophosphatasia was made.

## Management

SK's exfoliated teeth were submitted for histopathological analysis and the patient was referred to a paediatric endocrinologist: bone and mineral physician at the Children's Hospital at Westmead for further medical investigation. SK was referred for further biochemical testing including blood (full blood count with differential white cell count, C-reactive protein, liver function test, magnesium, urea and electrolytes, thyroid function test, genetic testing, and 25-hydroxy vitamin D), radiography of wrist and knee, and urine. Repeat blood analysis confirmed low levels of ALP.

Hard tissue microscopic findings: were consistent with the diagnosis of hypophosphatasia lacking cellular or acellular cementum overlying the dentine. ***(Figure 3+4)*** Hard tissue sections at high and low magnifications.)

## Follow up

Following medical review, a definitive diagnosis Odontohypophosphatasia (OHP) was made and long term management including recall with the Endocrinology team and regular dental follow up was arranged for the family.

## Recall dental appointment 2: Age 24 months old

SK attended for a dental review 6 months later. Her parents did not express concerns with SK's verbal communication, ability to eat or oral hygiene regime. SK was reportedly meeting her developmental milestones appropriately and her medical history was unremarkable.

On examination, SK appeared fit and well with an alert and happy disposition. Intraoral findings included healthy soft tissues, erupting primary teeth (upper and lower primary canines) and increasing mobility with teeth 61 and 51 (Grade II). The gingival margins around 61 and 51 were receded with resultant increased clinical crown lengths.

Teeth 82 and 72 appeared to have moved distally and also exhibited Grade I mobility. All primary first molar teeth were clinically sound. As such, no treatment was deemed necessary, however SK's parents were made aware of the possibility of further tooth loss. The importance of oral hygiene was emphasized and a follow up 3-month review was advised.

## Discussion

*Aetiology:*

This rare autosomal dominant and recessive condition is caused by mutations in the liver/bone/kidney ALP gene that encodes tissue non-specific alkaline phosphatase

*Figure 4: Hard tissue sections at low magnification*

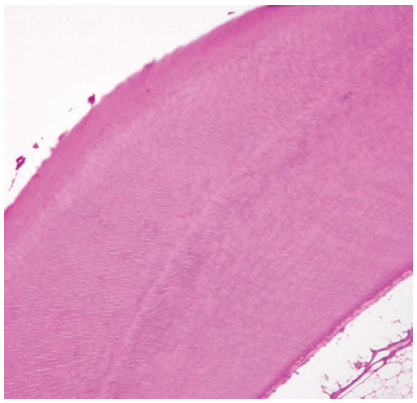


*Figure 5: Hard tissue sections at high magnification*

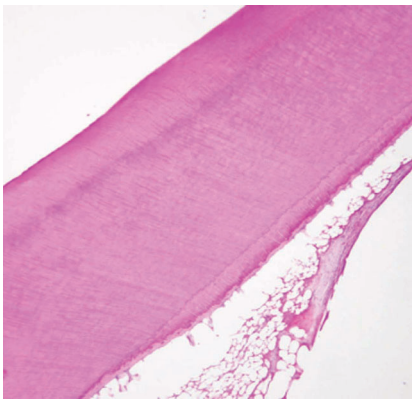


*Figure 6: Extraoral photograph at follow up*

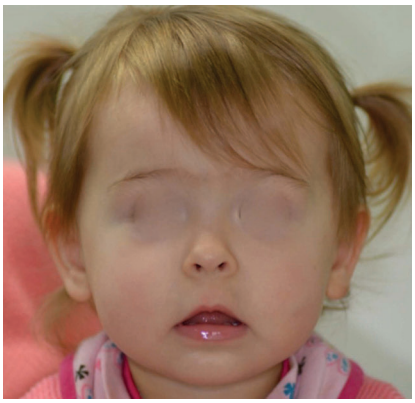


*Figure 7: Intraoral photograph at follow up*

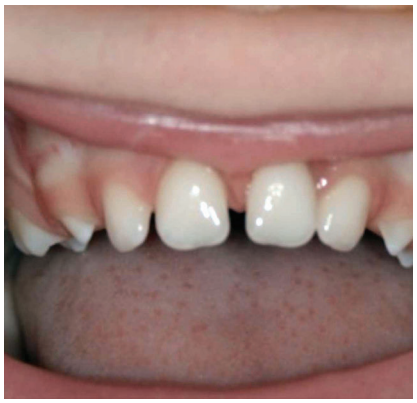
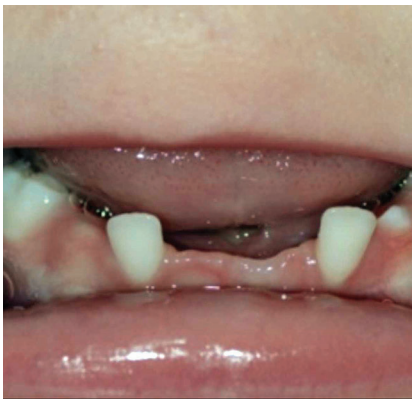


*Figure 8: Intraoral photograph at follow up*



(TNSALP). TNSALP has an important role cleaving phosphate from various enzymes namely pyriodoxxal-5-phosphate (PLP), phophethanolamine (PEA) and inorganic pyrophosphate (PPi). Reduced ALP activity results in defective mineralization of bone, causing widespread skeletal manifestations.

*Dental manifestations:*

The most significant dental finding is the premature loss of primary teeth. This clinical finding is often the first key signs of HP and is thought to result from agenesis or hypoplasia of cementum. Exfoliated teeth are often lost before resorption occurs presenting as fully rooted teeth (crown and root) and most commonly affect anterior teeth [5]. These dental manifestations are generally associated with milder forms of HP namely, infantile, childhood and OPH. However, severe forms of HP have resulted in posterior teeth exfoliating prematurely [6]. Radiographical features of the dentition includes reduced alveolar bone and enlarged pulp chambers and root canals [3]. Some authors report HP being associated with severe dental caries, enamel hypoplasia, bulbous crowns, delayed eruptions and delayed dentine formation [7, 3, 8]. In the permanent dentition, similar dental features have also been reported in the literature [9].

*Diagnosis:*

A referral to a paediatrician or paediatric endocrinologist for further investigation is indicated given the potential for systemic causes. The earlier the diagnosis of HP allows improved support and the aid of genetic counselling for families and patients involved. A definitive diagnosis requires a number of patient and family history details, extra oral findings such as height, weight and gait and intraoral clinical examination supported by radiographs [9].

Intra oral specific examination includes: dental hard and soft tissues, oral hygiene, tooth mobility and periodontal status as well as any abnormalities in the eruption pattern/dental morphology.

*Dental follow up:*

A dental management protocol is not well documented in the literature. Patients with HP display such a varied clinical presentation that dental care must be tailored to each individual. The ultimate aim for these patients is establishing a well – executed prevention program to ultimately reduce the risk of dental disease and tooth loss during all stages of life. The prevention of periodontal disease progression requires careful monitoring, as changes in progression may be indicative of systemic involvement. For young patients, it is recommended that a specialist paediatric dental team facilitate regular dental health reviews, as it is not uncommon to require a multidisciplinary team approach in the management of these patients.

*Prevention care:*

Establishing rigorous oral hygiene

- Using preventative hygiene measures both at home and in the dental clinic
- Regular periodontal health reviews

Reconstructive care:

- Replacement of permanent exfoliated teeth, removable appliance if required.

Maintenance phase:

- Regular dental reviews six monthly
- Referral to other dental specialities for multidisciplinary care.

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# Dental management of the Type 1 Diabetic Child

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## Introduction

The maintenance of oral health in the diabetic child provides many challenges. Diabetic children have an association with poorer oral health, particularly periodontal health. Children with diabetes mellitus require excellent dental management to maintain long-term oral health and minimise complications. Conversely, if the child is poorly managed the long-term consequences can be substantial for both their oral and general health. This review will examine the dental management of a Type 1 diabetic mellitus (T1DM) child.

The pancreas is crucial in maintaining body homeostasis and can be broadly divided into the endocrine and exocrine pancreas. The endocrine pancreas is primarily centred on the production of insulin and glucagon. In addition, somatostatin, a hormone involved in digestion and neurological function, and pancreatic polypeptide, a hormone involved in pancreatic self-regulation and hepatic glycogen levels, are produced and released by the pancreas. The exocrine pancreas produces digestive enzymes that assist in the digestion of carbohydrates, proteins and lipids.

Glucose is the most important cellular energy source derived from the diet and is transported in the blood. In healthy children, the normal blood glucose level (BGL) is between 4 – 6 millimoles per litre of blood (mmol/L) reaching up to 7.8 mmol/L 2 hours after eating (1). There is some conjecture as to what a healthy range is in a diabetic child, however, target levels recommended before meals are 4 – 7 mmol/L and 2 hours after meals, 5 – 9 mmol/L (1). A healthy range in a diabetic child is dependent on factors, such as the age of the child, insulin regimen and time since previous meal. (2).

Failure to adequately control BGL can result in episodes of hyperglycaemia or hypoglycaemia. Several hormones are involved in the regulation of BGL and these include: insulin, glucagon, adrenaline, thyroxine, cortisol and growth hormone. Dysregulation of glucose levels can have profound acute, medium and long term impacts on the health of a child (3).

## Diabetes Mellitus

Diabetes mellitus (DM) is a metabolic disease that is a consequence of an absolute or relative lack of insulin. Insulin is produced in the beta cells of the pancreas (islets of Langerhans) and is released in the presence of glucose. It serves a crucial role in many cells and organs and is heavily involved in the metabolism of lipids, carbohydrates and proteins. The net action of insulin is to reduce BGL with an increase in energy storage via adipose tissue, glycogen storage in the liver and muscle anabolism (4-6).

The three main types of diabetes are T1DM, Type 2 (T2DM) and gestational. The types relevant to paediatric oral care are T1DM and T2DM. T1DM is due to an absolute lack of insulin. The pathogenesis of T1DM is hypothesised to be due to exposure to an environmental antigen in a genetically susceptible individual. This results in autoimmune-mediated destruction of the beta cells in the islets of Langerhans. This process occurs over many months or years with the child asymptomatic until there is a large loss of functioning beta cells (7).

T2DM is due to a progressive failure of insulin production, a down regulation of insulin receptors and impairment of receptor function. Symptoms tend to develop slowly over time with risk factors including a poor diet that is high in sugars and trans fats, a lack of exercise, obesity, maternal gestational diabetes, genetic predisposition and ethnicity (2-4, 8, 9).

Gestational diabetes affects pregnant mothers and is best defined as an insulin resistant state resulting in an elevated risk of developing T2DM later in life (3, 10)

T1DM is the principal form of DM the paediatric dentist will encounter. In the general population it constitutes approximately 10% of DM cases. In the paediatric population it constitutes 94.5% of all DM diagnosis for children aged 0 – 14 years (11). Peak diagnosis occurs between 10 - 14 years-of-age, however, can occur at any age. In 2013 the prevalence rates for Australian children aged 0 – 14 years was approximately 140 per 100,000 population with an incidence of 25 per 100,000 people (12).

T2DM constitutes approximately 85% of all DM cases in the general population (11). In the paediatric population it is extremely rare under the age of 10; constituting 17% of DM cases in 10 – 19 year-olds. In 2012, the incidence for 10 – 14 year-olds was 2.6 per 100,000 with a prevalence of 0.01%. For 15 – 19 year olds an incidence of 8 per 100,000 was recorded and a prevalence of 0.04%. No significant increase in incidence was noted over the preceding 10 years, however, the risk factors for T2DM, inactivity and obesity, are steadily increasing (8).

The diagnosis of T1DM is made commonly on clinical presentation followed by confirmation from blood glucose testing. The classic signs and symptoms include polydipsia, polyuria, weight loss with hyperglycaemia and ketonuria. Diabetic ketoacidosis and incidental discovery can also lead to a diagnosis (1, 3, 7, 13, 14).

Blood glucose testing is used to support clinical suspicions. Four tests are possible and include a fasting blood glucose >7 mmol/L, a random venous blood glucose of >11.1 mmol/L, a blood glucose >11.1mmol/L following an oral glucose tolerance test or a glycated haemoglobin (HbA1c) >6.5% (>7.7 mmol/L). The HbA1C test measures the percent of haemoglobin A bound to glucose via non-enzymatic glycation. It gives the practitioner an average BGL over a 10 – 12 week assessment. This is particularly useful in long-term management and treatment of the child to assess compliance with treatment regimens (7).

T1DM has a moderate-to-strong familial history as evidenced by 0.4% spontaneous prevalence compared to up to a 50% prevalence with an affected monozygotic twin (7). Furthermore, there is an association with genetic syndromes (Down syndrome, Turner syndrome, Prada-Willi syndrome, Friedreich ataxia), cystic fibrosis and hereditary hemochromatosis (diseases of the exocrine pancreas), coeliac disease and other endocrinopathies involving anti-insulin hormones (cortisol, adrenaline, growth hormone and thyroxine) (15).

The complications of poorly controlled DM are multi-system in nature and involve acute medical emergencies up to long-term complications such as limb amputations. Acute complications can include hypoglycaemia, hyperglycaemia and diabetic ketoacidosis. The impact on growth and development needs to be monitored as poorly controlled T1DM can lead to poor growth and weight gain (16).

Long-term complications of poorly controlled T1DM include nephropathy, retinopathy, neuropathy and cardiovascular disease. These occur secondarily to vascular complications and typically

become apparent in adulthood (16). The pathogenesis is incompletely understood, however, is thought to be due to episodes of elevated BGL resulting in osmotic stress in insulin-independent tissues. Excess glucose is converted into sorbitol, which can glycate proteins (collagen and elastin) forming advanced glycation end-products (AGEs). This decreases elasticity, healing and strength as well as stimulating low grade inflammation. Hyperglycaemia also results in an increase in reactive oxidative species and a reduction in neutrophil function (4, 5, 16, 17).

## Medical Treatment of DM

Children and adolescents can be difficult to medically manage due to their stage of development and unpredictability in maintaining consistent dietary intake, insulin administration and exercise (7).

Treatment programs focus strongly on diet, exercise and insulin administration with self/parental monitoring. Poor glycaemic control, as evidenced by hyperglycaemia following meals, places the child at a significantly elevated risk of long term complications. A HbA1C within the target range of <7% (8.5mmol/L) indicates good control (7, 16, 18, 19).

Insulin is the mainstay of treatment in T1DM. Insulin is available in rapid, short, intermediate and long acting forms and is commonly administered via subcutaneous injections (3, 20). Rapid acting insulin, such as Aspart® and Lispro®, are commonly administered 5 - 15 min before meals. They have an onset of 15 min, a peak of 1 hour and duration of 2 - 3 hours. Short acting insulin, such as Humulin® R, is commonly administered 20 – 30 min before meals. They have 30 min to 1 hour onset, a peak between 2 – 4 hours and duration of 4 – 6 hours. The dosage will vary depending upon the carbohydrate content of the meal and the BGL. Intermediate acting insulin includes forms such as Humulin® NPH and is administered two to three times-a-day, in combination with longer acting insulin, and will provide some cover for meals. They have an onset of 30 min to 1 hour, a peak of 4 - 6 hours and duration of 8 – 16 hours. Long acting insulin, such as glargine, is administered once or twice-a-day, with an onset of 30 min to 1 hour, no peak and a duration of 24 - 26 hours. The long-acting insulins are commonly used to provide basal insulin levels (19).

There is a range of regimens that can be implemented. The two most commonly used regimens are either a twice-daily injection of a mixture of short and intermediate insulin (conventional). This is performed in children under 10 years due to their inability to properly monitor and administer. The second regimen involves an intensive protocol with nightly long-acting basal insulin and three infusions of rapid-acting insulin pre-meals. The regime offers greater flexibility than conventional therapy but requires a child's independence for drug administration and monitoring. The insulin pump is classified as an intensive regime. It administers a continuous basal dose of rapid acting insulin with an increase (bolus dose) pre-meal. For the insulin pump, continuous glucose monitoring is required with adjustments accordingly (20). The diabetes control and complications trial conclusively showed that good glycaemic control delays the onset and progression of complications of T1DM (21).

## Oral complications of T1DM

Numerous studies have examined the role of T1DM in oral complications. These studies, however, focus on the adult population primarily. Studies pertaining to T1DM and the oral health of children are limited (22). Furthermore, the impact of T1DM on oral health is complicated and data commonly contradictory (14, 22, 23).

Gingivitis and Periodontitis: Periodontal diseases are the best-documented oral complication of T1DM and are due to the exaggerated inflammatory response, increased tissue destruction and poor neutrophil function (24, 25). This is mediated by AGEs and oxidative stress (14). The effect of periodontitis on T1DM is putatively due to the production of pro-inflammatory cytokines that antagonise insulin. There is evidence that periodontal therapy improves glycaemic control via a reduction in HbA1c (26).

Lalla and colleagues examined 700 children between 6 - 18 years with 350 diagnosed with T1DM and 350 healthy individuals. They noted statistically higher plaque levels, gingivitis and greater attachment loss in T1DM children. A demonstrated association between diabetes and an increased risk of periodontal destruction in children and adolescents was determined (18). The majority of studies have also indicated poorer periodontal health with higher plaque accumulations and gingivitis in T1DM children compared to healthy controls (23, 27-33). Limited evidence for no difference or better periodontal health and plaque control in T1DM children exists (23, 34, 35). Due to the evidence indicating an association between poor periodontal health and T1DM, dental management of T1DM children requires early identification of gingivitis, gingival bleeding and plaque, excellent oral hygiene and a more frequent recall regimen (6, 18, 22-24).

Dental Caries: Caries experience for children and adolescents with T1DM is highly variable in the literature. This is as authors have reported an increase in the caries experience (29, 32, 36, 37), a decrease (27, 38, 39) and no difference (28, 40-42). In case-control studies in the permanent dentition, an increase in the caries experience, as measured by mean DMFT or DMFS, was reported for individuals with DM compared to healthy controls (29, 32, 36, 37). The mean DMFT range in T1DM children was 1.8 - 11.2 compared to 0.6 - 9.6 in healthy children (32, 36, 37). This difference was statistically significant ($p < 0.05$) (29, 32, 36, 37). Other authors reported a statistically significant lower caries experience in mean DMFT for T1DM children (1.7 - 4.6) compared to healthy children (5.5 - 6.4) (27, 38). One author reported a statistically significant mean DMFS for T1DM children of 23.0 compared to 27.4 for healthy children (39). In summary, these studies varied significantly in design, geographical region, sample size, age of participants and statistical analysis (23). Furthermore, most studies recruited children seeking treatment in dental clinics and therefore were not representative of the general population (23).

In the primary dentition, there was no recorded increase in the caries experience, as measured by dmft or dmfs, of T1DM children compared to healthy controls (27, 28, 40, 42). In longitudinal studies, no increase in the caries experience over the study period was noted (43-45) when compared to healthy controls. It has been concluded that there is no direct association between T1DM and caries experience (23, 44). Consequently, it can be hypothesised that the link between DM and dental caries is complex and multifactorial, with many risk factors that need to be considered (14, 23).

Salivary Dysfunction: Variables related to salivary function analysed included pH, flow rate and buffering capacity. There was variable and contradictory evidence for salivary characteristics. Further complicating comparison, there was significant differences in study design, variables measured, age of population and geographical region. Some authors noted a statistically significant lower salivary pH for T1DM children compared to healthy controls with a range between pH 6 – 7 and pH 6.9 – 7.1 respectively (30, 32, 46). One author noted no change

in pH (41). A statistically significant lower unstimulated saliva flow rate was reported in T1DM children (0.2 - 0.7 mL/min) compared to healthy controls (0.3 - 1.1 mL/min) (32, 39, 42, 44, 46-48). Conversely, two authors noted no difference in flow rate (30, 41). The reduced flow rate in T1DM is putatively due to dehydration secondary to hyperglycaemia and neuropathy affecting salivary gland function (32, 39, 42, 44, 46-48). A decreased buffering capacity was found by one author (30) with another reporting no difference (41). In conclusion, a greater number of authors found a reduction in unstimulated salivary flow rates and salivary pH than the contrary position.

Oral Mucosal Diseases and other oral Infections: No literature in children regarding the impact of T1DM and oral mucosal diseases was found. In adults with T1DM there has been a more consistent finding of increased prevalence of oral candidiasis than healthy subjects (14, 49, 50).

Orthodontic considerations: Orthodontic treatment can be conducted safely in children with good oral hygiene and well controlled DM. Due to the risks of periodontal issues, poorly controlled T1DM children with a HbA1C >9%, are contraindicated for orthodontic treatment. Children with well controlled T1DM should be treated as if they already have periodontal disease, with an increased vigilance and review regimen. Diabetes-related microangiopathy has been postulated to compromise periapical vascular supply resulting in pulpal symptoms and potentially loss of vitality. Consequently, orthodontic forces should be light and frequent sensibility testing should be conducted (13, 51).

## Dental Management of a Type 1 DM Child

Dental management of T1DM requires a methodical and careful approach. Children who are poorly managed in the dental setting are at higher risk for an acute medical emergency. Secondarily, excellent dental management can avoid unnecessarily stressful situations from developing and help prevent and mitigate complications that can develop as a consequence of this disease.

Following a comprehensive dental assessment, it may be decided that procedural dental care is required. Prior to performing dental treatment the medical history of the child needs to be established. The treating paediatric endocrinologist needs to be contacted to gain this information. This includes age at diagnosis, the history of diabetic complications (frequency of hypo- or hyperglycaemic episodes) and hospitalisations, determining their insulin regimen, the most recent HbA1C measurement and most importantly their current diabetic control (6, 24, 25, 52).

For poorly controlled T1DM children any elective procedure is to be delayed until adequate control has been established and following clearance from the managing paediatric endocrinologist. For children requiring emergency treatment, referral to a hospital setting, such as a specialist children's hospital, is required. Here the dental team will liaise with the paediatric endocrinology team to manage the child appropriately (52, 53).

For children who are well-controlled diabetics, there are several recommendations to enable treatment to be performed safely and confidently in an outpatient setting. Procedures should be scheduled early in the morning with normal insulin and meal regimens. Prior to conducting treatment measurement of BGL is recommended, with an ideal BGL between 4 - 7 mmol/L. If the BGL is <4 mmol/L readily absorbed oral carbohydrates should be given followed by a re-measurement prior to commencing treatment (3, 6, 24, 25). If BGL are noted to be >10 mmol/L treatment would not be recommended at this concentration without consultation with the endocrinologist. For prolonged dental procedures re-measurement of BGLs throughout the procedure may be indicated (24, 53).

Effective pain relief is required during procedures and post-operatively in T1DM children to help reduce fluctuations in BGLs. Nitrous oxide anxiolysis and local anaesthetic with adrenaline are safe to use and will help reduce anxiety which may increase their pain threshold. Due to the increased risk of infection after significant surgery, or those children at high risk of oral infections, antibiotics should be considered. Post-operatively, the total caloric intake of the patient, including the fat-carbohydrate:protein ratio, needs to be maintained at pre-surgical levels to allow proper glycaemic control. If this is not possible due to the dental procedure, the paediatric endocrinologist must be informed as dietary and insulin dosage modifications may need to be made (6, 24, 25).

Children with T1DM are classified as American Society of Anesthesiologists (ASA) level 3 (54). If a general anaesthetic is required, referral to a hospital that is experienced in dealing with diabetic children is required (52). Care for the child will be coordinated by the paediatric dentist with the anaesthetist and paediatric endocrinologist. These children should be first on a morning list. For shorter procedures (less than 2 hours) the child is routinely admitted on the morning of the GA and discharged that afternoon after oral intake is restored (53).

For complex or prolonged procedures, which may impact on oral intake following the procedure, the paediatric endocrinologist may recommend hospitalisation the day before GA and post-operatively to monitor and adjust the insulin regimen. The child will be discharged when adequate oral intake is restored to the satisfaction of the paediatric endocrinology team (52, 53).

In an out-patient setting, medical emergencies can occur, hence, the dentist must be competent in basic life support (BLS) and CPR. Rapid identification and timely management is critical to avoid serious complications. The primary concern in T1DM children is a hypoglycaemic episode. Signs and symptoms of hypoglycaemia include hunger, weakness, sweating, anxiety, irritability and tachycardia (3, 6, 55).

## Summary

T1DM is a metabolic disease that the public and dental practitioners must be aware of. Its prevalence in the paediatric population requires excellent identification and management with the dental team serving a crucial role in the child's care. The deleterious impact of T1DM on periodontal health has been noted. Associations with other oral conditions, such as carious lesions, salivary characteristics and oral mucosal diseases is less clear. Regardless, ensuring children with T1DM have excellent oral health is crucial in maintaining good glycaemic control and preventing the development of short and long-term complications of this metabolic disease.

The dental practitioner treating T1DM children must have a sound knowledge of the disease, be in communication with the paediatric endocrinologist and ensure excellent treatment planning. This will ensure the treatment of the child with T1DM can be performed safely, confidently and comprehensively.

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# Up Coming *Events*

### 13 March 2017
**ANZSPD (Qld) Dinner Meeting**
Victoria Park Golf Course, Herston QLD
go.65@optusnet.com.au

### 14 March 2017
**ANZSPD (NSW) Dinner Meeting**
Mantra Chatswood, NSW
anzspd.nsw@gmail.com

### 17 March 2017
**ANZSPD(WA) Partner's Evening**
Guest speaker: Mike Wood
Nunzio's, Fremantle WA.
anzspdwa@gmail.com

### 26-27 March 2017
**RK Hall Lecture Series 2017**
Auckland, New Zealand
http://bit.ly/28KASeo

### 8 May 2017
**ANZSPD(Qld) Dinner Meeting**
Victoria Park Golf Course, Herston QLD
go.65@optusnet.com.au

### 9 May 2017
**ANZSPD(SA) Fluoride – friend or foe?**
Hackney Hotel & Function Centre, SA
gwendolyn.huang@gmail.com

### 17-21 May 2017
**ADA 37th Australian Dental Congress**
Melbourne, Australia
http://www.ada.org.au/Congress

### 21-22 July 2017
**ANZSPD(WA) Bunker Bay Mid-Winter Meeting**
Pullman Bunker Bay Resort, Naturaliste, WA
anzspdwa@gmail.com

### 4-7 October 2017
**IAPD 26th Congress**
Santiago, Chile
www.iapd2017.com

### 15-18 February 2018
**ANZSPD Biennial Conference**
Seaworld Resort
Gold Coast, Australia.

## Australia and New Zealand Society of Paediatric Dentistry
## www.anzspd.org.au

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